

Amendments to the Claims:

This listing of claims replaces all prior versions and listings of claims in the application:

Listing of Claims:

1. (Currently Amended) A method of identifying a candidate therapeutic compound for treating facioscapulohumeral muscular dystrophy (FSHD), the method comprising:
 - (a) providing a first D4Z4 binding element comprising at least one nucleic acid sequence of SEQ ID NO: 21 or at least one nucleic acid sequence of SEQ ID NO: 23 ~~DNA comprising one or more 4q35 D4Z4 repeats~~;
 - (b) contacting the first D4Z4 binding element with a test compound;
 - (c) determining whether the test compound interacts with the first D4Z4 binding element;
 - (d) selecting the test compound if the test compound interacts with the first D4Z4 binding element;
 - (e) providing a cell comprising a second D4Z4 binding element that comprises at least one nucleic acid sequence of SEQ ID NO: 21 or at least one nucleic acid sequence of SEQ ID NO: 23, and is operably linked to a FSHD region gene 1 (FRG1);
 - (f) contacting the cell with the selected test compound;
 - (g) determining the level of FRG1 expression in the cell in the presence of the test compound;
 - (h) comparing the level of FRG1 expression in the cell in the presence of the test compound to a reference level of FRG1, and
 - (i) selecting the test compound as a candidate therapeutic compound for treating FSHD if the level of FRG1 expression in the cell in the presence of the test compound is reduced as compared to the reference level of FRG1.

2. (Currently Amended) The method of claim 1, wherein the second D4Z4 binding element is cell-comprises an endogenous D4Z4 binding element and ~~operably-linked FSHD region gene 1 (FRG1).~~

3.-4. (Cancelled)

5. (Previously presented) The method of claim 1, wherein the cell is a muscle cell.

6. (Previously presented) The method of claim 1, wherein the cell is from a subject that has FSHD.

7. (Currently amended) The method of claim 1, wherein the interaction is the binding of the test compound to the first D4Z4 binding element.

8. - 9. (Cancelled)

10. (Withdrawn, Currently amended) A method of identifying a candidate therapeutic compound for treating facioscapulohumeral muscular dystrophy (FSHD), the method comprising:

- (a) providing a first D4Z4 binding element (DBE) comprising ~~DNA comprising at least one nucleic acid sequence of SEQ ID NO: 21 or at least one nucleic acid sequence of SEQ ID NO:23~~ 4q35-D4Z4 repeats;
- (b) contacting the first D4Z4 binding element with a test compound;
- (c) determining whether the test compound interacts with the first D4Z4 binding element; and
- (d) selecting the test compound if the test compound interacts with the first DBE D4Z4 binding element;
- (e) providing a second D4Z4 binding element comprising at least one nucleic acid sequence of SEQ ID NO: 21 or at least one nucleic acid sequence of SEQ ID NO:23 ~~DNA comprising 4q35-D4Z4 repeats~~ and a D4Z4 recognition complex

(DRC) or component thereof under conditions such that the second DBE and the DRC or component thereof can bind to each other;

(f) contacting the second D4Z4 binding element and DRC or component thereof with the selected test compound; and

(g) determining whether the test compound affects the binding of the second D4Z4 binding element to the DRC or component thereof,

wherein an increase in binding between the second D4Z4 binding element and the DRC or component thereof in the presence of the test compound indicates that the test compound is a candidate compound for treating FSHD.

11. (Withdrawn) The method of claim 10, wherein the DRC component is YinYang 1 (YY1), High Mobility Group Box 2 (HMGB2), or nucleolin.

12. - 20. (Cancelled)

21. (Currently amended) A method of identifying a candidate therapeutic compound for treating facioscapulohumeral muscular dystrophy (FSHD), the method comprising:

(a) providing a cell expressing a D4Z4 binding element reporter construct comprising at least one nucleic acid sequence of SEQ ID NO: 21 or at least one nucleic acid sequence of SEQ ID NO: 23 ~~DNA comprising 4q35 D4Z4 repeats,~~ operably linked to a promoter and a reporter gene;

(b) contacting the cell with a test compound;

(c) determining a level of expressing of the reporter construct in the presence of the test compound;

(d) comparing the level of expression of the reporter construct in the presence of the test compound to a reference representing a level of expression in the absence of the test compound; and

(e) selecting the test compound as a candidate therapeutic compound for treating FSHD if the level of expression of the reporter construct in the presence of the test compound is reduced as compared to the reference.

22. (Cancelled)

23. (Currently amended) The method of claim 21, wherein the reporter construct comprises one, two or six ~~minimal~~ D4Z4 binding elements.